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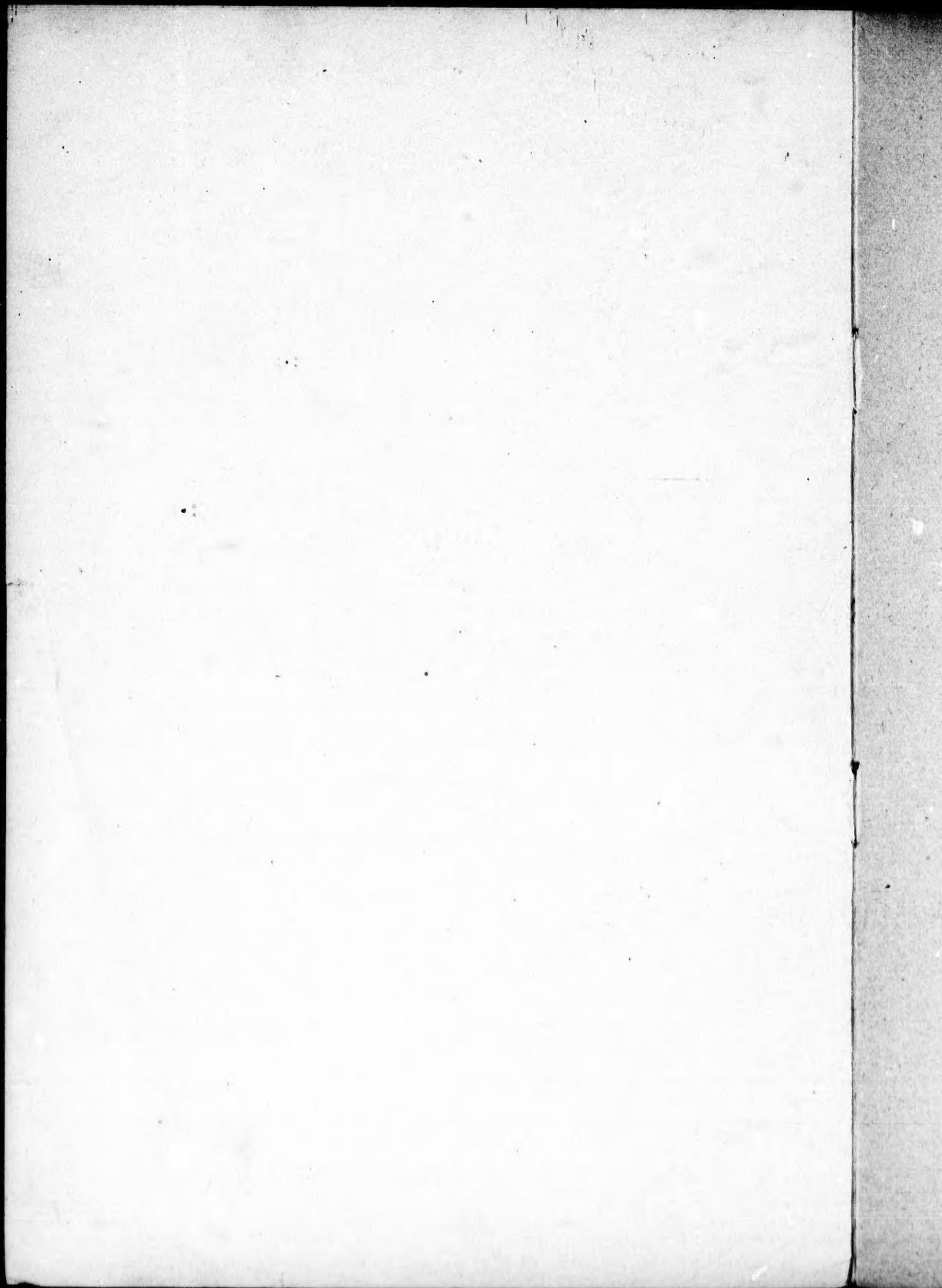
ALBINISM.

BY

W. H. DALPÉ, B.A., MONTREAL

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ALBINISM.¹

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W. H. DALPÉ, B.A., MONTREAL,

Definition.—Albinism is usually defined as a hereditary condition marked by a complete absence of pigment in the iris, skin and hair. However correct this statement is, a long residence with albinos in this Province and in New England, has led me to associate with this condition certain functional and organic alterations in the skin and appendages, and about which I will have to say a few words.

Albino means white and was a term used by the Portuguese to designate the white negroes whom they met on the Western Coast of Africa; by extension it is used to designate any pigmentless individual.

Occurrence.—Albinism is an affection of a world-wide distribution and not limited to the dark races as was surmised. In the Medical Literature, of Europe and America, several accounts have been given, none better or more tersely written than that found in Morrow's Handbook of skin diseases, by one of our best dermatologists.

This affection is not peculiar to man but similar conditions are found to exist among the animals, birds and even insects.

CHARACTERISTICS.

Eyes.—The irides are usually perfectly pigmentless. In many, these transparent perforated screens take on a pinkish or reddish hue from the capillaries of the fundus. Balmanno Squire reported a case, which he calls atypical and in whom the irides were dark blue *Lancet*, Feb. 1895. Zeimssen recognising that such blue irides do occur claims this is due to some phenomenon of interference of light. They have appeared to me, on more than one occasion, very much chameleon-like, the most constant colours being the white and the pink.

Consequent upon this pigmentless condition of the irides, the camera obscura of the eye is changed to a camera lucida, and the image falling on the retina, being more diffuse, loses proportionately of its clearness.

Again as the sensitive eye-plates of the retina are ill-protected, nystagmus,² photophobia,³ coloboma, (Wm. Geo. Sym, Edin.), and oscillatory movements of the eyes may be induced (A. J. Balmanno

¹ Prize Essay McGill Medical Society, 1897.

² "Nystagmus" and "oscillatory movements" though in some respects analogous, are not necessarily identical.

³ Usually congenital, and hence it may be doubted that non-pigmented irides hold any causative relations to it.

Squire.) But it must not be inferred that all albinos will evince what Prof. Shepherd happily terms "the screwing of the eyelids" under a strong light; neither are they all weak-eyed; Squire's case could read perfectly in daylight and in the numerous cases which it was my good fortune to see, although their vision was keener at night, yet by daylight they saw perfectly well, neither did I see any resort to the wearing of glasses. (*Ab albinismi causa.*)

Hair.—The hair is possessed of a greater or less degree of whiteness, at times immaculate, at times flaxen, whitish yellow, always exceedingly fine and of a silky lustre, sparse, leaving the brow and nucha well denuded and apt to leave the corona bald in men.

This condition especially in women, often, if not always, demands the wearing of false hair. In children their hoary denuded head in the presence of the other conditions makes a picture most sad to behold.

Beards were not to be seen among the men in my cases, yet some would see fit to encourage the tonsorial artists, at long intervals, by requesting the removal of what seemed to me more like down than beard. What this would come to, if left unmolested, I am unable to say, other than it must of necessity be a caricature of the genuine article.

In the axillæ and over the pubes, the growth of hair was seen to be more vigorous, and the hair yellowish-white, curly, more brittle—a condition which was no doubt due to the hyperidrosis of the part.

The Skin is also devoid of pigment, but it may acquire a rose colour from the underlying vessels, in places where the integument is thin as over the cheeks and the dorsum of the hands. This gives the albinos a pale, sallow, sickly appearance; extreme anæmia may appear to exist.

The skin is soft to the touch, dry, in some places very thin. Yet in the portions of the body subjected to pressure as the surface of the hands or the soles of the feet the skin was granular, rough, full of rugæ not only in working people but in individuals not inured to hard labour and in whom, presumably, this condition could not be due to this cause.

In the normal, the pigment occurs between the cuticle and the cutis vera; here seemingly an epiblastic derivative has one of its functions absent, whilst in the eye some mesodermic derivative is at fault.

The Nails. I am now going to speak of a condition which was found to exist concurrently with those already named, in a great number of cases. I refer to a particular conformation of the nails; this was so characteristic, so general in the cases that came under my

notice, that a diagnosis might easily have been made by a single exhibition of the finger tips. The nail-base or matrix does not reach to the end of the finger; is only about half as long as in other people, and is made up of a semi-circle directed upwards, and a triangle looking down towards the finger tips. In the normal the nail matrix is more circular or oval.



Normal.



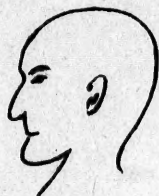
Albino.

Moreover the tip of the matrix was raised and the nail given an arched direction so that on a longitudinal section it would look very much like a claw (onychogryphosis) a resemblance which was very much increased by a pathological overgrowth or hyperonychia, rendering them clumsy, thick, brittle, useless for scratching or opening a knife blade. Albinos in whom this condition of the nails is found usually either hide them in other people's presence, or, to improve their appearance, trim them very short, close to the stumpy base or matrix.

Why such a condition should occur concurrently with albinism, I am not in a measure to explain. The points which especially struck me were the congenital nature of this gryphotic condition; its frequent if not absolute association with the pigmentless condition; the involvement of all the nails at the same time. The condition which most resembles that just referred to, is probably onychomycosis due to a fungus or a somewhat similar one due to *tinea favosa* or *trichophyton tonsurans*. Here the nails are brittle, frayed out, intersected by furrows, opaque, grayish or yellowish-white, and are lifted up and gryphotic, disfigured; the matrix being implicated, changes of growth are also present and the matrix may be painful, swollen, suppurative, (Zeimssen). This author also includes as possible etiological factors, scrofulosis, cachexia, oligæmia, and Billroth also mentions moist gangrene. Again in scleroderma, sclerodactylism may simulate onychogryphosis. But in all these changes there would be a lack of conformity to the same type, an irregularity in distribution, a history of infection or a progressive condition, in some suppuration, in others extreme deformity of the finger tips. Moreover these would not be congenital as was that condition which I observed in my albinos.

The teeth were also found to be hypoplastic or stunted in a variable

but always in a marked degree ; they were squarish, and stood apart; this gave my albinos the appearance of old people, the mouth receding with its stunted teeth, standing in marked contrast to the prominent chin and nose.



Here there was no gouging of the middle incisors as is apt to occur in congenital lues, nor was there any other evidence of this virus in the system ; however, in one family I was given to understand that a leutic taint existed. Their teeth did not seem to tend to early decay.

Bromidrosis. A fetid perspiration has long been recognised in some pathological conditions ; thus scrofula, rheumatism, lues, the parturient state have all been associated with a particular emanation. However in some it cannot be connected with any morbid conditions. In such albinos and sons of albinos with whom I played, walked, slept, a peculiar, penetrating, not absolutely unpleasant odour was at once perceptible, which forced ablutions could only dispel for a limited time. Moreover, it was asserted to me that this was increased in females at the menstrual period, but whether it was complementary or vicarious my informers were unable to state. *Excepi cum grano salis.* Zeimssen says the functions of the skin are normal.

The albinos barring these physical differences do not seem to be an inferior class of people. The authors usually say they are of a weakly constitution, but Zeimssen says this is a rule with manifold exceptions. I have always been of opinion that this was more apparent than real. The general cachectic appearance might easily be attributed to the general pigmentless condition of the skin and hair, the weak eyes and receding mouth. Many, I have found, to be surprisingly agile and strong. Several severe shakings which I got at school, by an albino, the seventeenth child of an albino mother, have forcibly convinced me that albinos are not physical degenerates. This particular boy, conscious of his own strength and endurance, was wont to afford us evidences of them by lifting steel rails and by running to a pump, a distance of 150 yards, in cold stormy mid-winter nights, bareheaded, barefooted, and with only thin underwear on, and there to fill his pitcher and to bring it back full. Nor was he ever sick as a conse-

quence. Neither was he intellectually deficient, but he now wears a clerical dress, with at least, much decorum and success.

In a family of which four generations have come under my observation, the representative of the first was a man of unusually ready wit; his son, though only five feet three inches in height, was a wiry little personality; in the third generation were observed four professional men and a fifth an albino, a sculptor.

Of course the albinos ordinarily are not all physically and intellectually the par of those just referred to. My albinos were for the most part long-lived, not phthisical; I have not yet come across a case of idiocy amongst them.

They are generally discriminated against when it is a question of giving or taking in marriage, and they, as a class, feel that a stigma attaches to them, whereby they are often rendered abashed, morose, melancholic. This has been noted by most authors. Menfolks usually marry others than albinos, but the women usually do not marry at all, probably for want of suitors, unless there be a large dowry in sight. Thus the increase amongst the albinos is not a very rapid one.

The sexual propensities are very highly developed in some if not in most and have been noted in the non-albino progeny of mixed unions. *Sed hoc non semper albinum est.*

Etiology.—Heredity is recognised as the most important factor in the causation of the disease. Thus some parents, immediate or remote are looked upon as the channel of transmission; the atavism may be so great that the cases may appear sporadic. Case of Balmanno Squire, the *Lancet*, Feb., 1895. The cases of Geo. Wm. Sym, Edin.

The children of an union between an albino and a normal individual are for the most part normal; rarely do we see the majority of them pigmentless. A child having a full development of hair may yet have a stunted growth of teeth or may even have a partial onychogryphosis, or again these may disappear from one generation and reappear in some members of the next to tell its tale of a lurking albinism.

As a second possible etiological factor some include some telluric conditions. Thus it is claimed that it occurs endemically in Loango or Lower Guinea. However, such telluric causation is usually not admitted. Beyond this we enter the realm of hypothesis. Are we to look for a cause within the part affected or to some anomaly in the ductless glands? No answer is at present forthcoming.

Classification.—Bärensprung classified pigment atrophies into *acquired* albinism or vitiligo, and *congenital* albinism which he further sub-divided into *universalis* and *partialis*. The first and the last he

misused, using the one for morphea and *sclérodémie en plaques*, the other for vitiligo. (Zeimssen.)

Vitiligo or leucopatia acquisita is a progressive pigment atrophy, which is regardless of sex, appears usually between the tenth and thirtieth year, may remain stationary or extend over the whole body or may even disappear spontaneously. It may run a symmetrical course on both sides of the body. The skin around these white spots usually becomes more pigmented. Näcke Lévy. It is often seen in cases of acquired syphilis. When it involves the scalp, canities result. *Berliner Klinische*, Feb., 1894. In the course of the diseases of the thyroid leucopatia acquisita may occur and by the total involvement of the skin may even simulate albinismus universalis. In Graves' disease, however, there would be tachycardia, an enlarged thyroid, venous pulsation, muscular tremors, and characteristic eye-changes.

In scleroderma either in the localised or the diffuse form, following hyperæmia and preceding induration, leucoderma often appears and often with it dystrophy and sclerodactylism. Here, given a very chronic case, symmetrically distributed gryphotic albinism might be very closely simulated. But in scleroderma, sensibility and secretion are lessened, there is bradycardia, and intellection is lowered.

In some cases of albinismus acquisitus partialis, the area of pigment atrophy may correspond to the area of supply of some nerve. From clinical notes I obtained two remarkable cases, one with a pigmentless spot corresponding to the hypogastric branch of the ilio-hypogastric, another with a white spot corresponding to the distribution of the subcutaneous *colli medius et inferiori*. I might also say that a somewhat similar auto-observation was made by myself several years ago.

Of congenital partial albinism, I need not say much. The colour merges gradually from the pigmentless spot into that of the healthy surrounding skin which is not more pigmented than elsewhere. In some canities may occur without a lack of pigment in the scalp. Stricker refers to a white lock of hair transmitted for six generations.

A pseudo-albinismus universalis congenitalis, I think, may be rightly said to occur. I am very much inclined to look upon Sym's four cases as such. The hair was extremely fair but not quite white; the irides of an exceedingly pale blue colour, not pink. It is not an uncommon thing to find individuals with very pale yellow or flaxen hair, eyebrows and ciliæ, and with almost pigmentless skin and irides, so that one almost feels as if they should be classed in a sub-variety

of albinism. These pseudo-albinos would be characterised by the fact that the lack of pigmentation is not so marked nor so persistent, by the gradual deepening in colour noticed in the hair as age advances by the appearing of freckles; here again beards will often grow, though not always; nor will there be usually gryphotic changes in the nails, nor hypoplasia of the teeth. Yet here, as in many other things, fast and hard rules are only good enough to be broken.